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Erythrodermic Dermatomyositis

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Erythrodermic Dermatomyositis

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Case Presentation:

Patient: 50 year-old Caucasian male.

History of Present Illness: This patient presented with a 15-week history of a rash over his entire body. The rash is associated with pruritus and edema. Prior treatments include prednisone, hydroxyzine, cetirizine, famotidine, and triamcinolone with some improvement. He reports odynophagia, unintentional 10 pound weight loss within the last 3-4 months, and occasional difficulty lifting both arms. He denied nausea, vomiting, headaches, changes in urinary patterns, and back pain.

Medical History/Surgical History: Hypothyroidism, sleep apnea, inguinal hernia repair

Family History: Hypertension, amyotrophic lateral sclerosis

Medications: Levothyroxine

Previous Treatment: Prednisone, hydroxyzine, cetirizine, famotidine, triamcinolone 0.1% ointment

Current Treatment: Prednisone, hydroxychloroquine, mycophenolate mofetil, clobetasol 0.05% ointment

Physical Examination: Generalized erythematous patches on the extensor surfaces of the upper extremities and most of the trunk. There is confluent erythema and edema of the lower extremities with sparing of the popliteal fossa and lateral feet. Scaly erythematous papules and patches on the dorsal hands with distinct flat-topped papules over the PIP and DIP joints and dorsal toes. Erythema and edema on face, ears, and the upper eyelids. There is hyperkeratosis and prominent telangiectatic vessels in the proximal nail folds with ragged cuticles. There is a palpable 1cm lymph node in the right axilla. Genitals are spared. No muscle weakness on initial exam.

Laboratory Data: (9/19/2014) Creatinine kinase 1148 (<351), AST 104 (<41), ALT 57 (<56), Aldolase 16.3 (1.5-8.1), ANA negative; (7/17/14) UA, urine porphyrins, serum complement, ACE, ESR, CRP, HLB-27 and creatine kinase WNL. Anti-thyroglobulin Ab 42 (0-40), anti-thyroid peroxidase Ab 337(0-34); (05/24/14) CMP, uric acid, RF, ESR, TSH and CBC WNL except lymphocyte count 0.79(1.2-4.8)

Studies: MRI brain, CT scan of head/neck, chest, and abdomen were negative for underlying malignancy (10/10/14). Colonoscopy, endoscopy (10/2/14), PFT (10/14/14), and left inguinal node biopsy (9/30/14) were negative for malignancy.

Biopsy: *CBL Path* (D14NY1-0234678, 08/15/14) Left 3rd finger dorsal DIP, right knee: Interface dermatitis. "Mild acanthosis and patchy parakeratoisis with sparse perivascular infiltrate". Left arm- Punch DIF: Granular deposition of IgG in epidermis and C3 in BMZ.

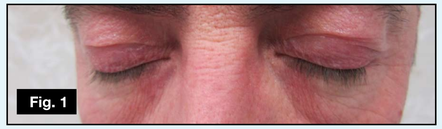

Figure 1: Heliotrope rash.

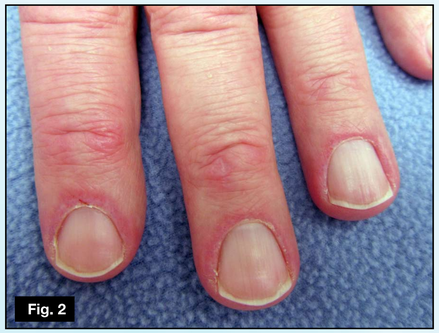

Figure 2: Erythema of the hands with Gottron's papules along the DIP joints and ragged cuticles.

Figure 3: Erythroderma of the neck and trunk.

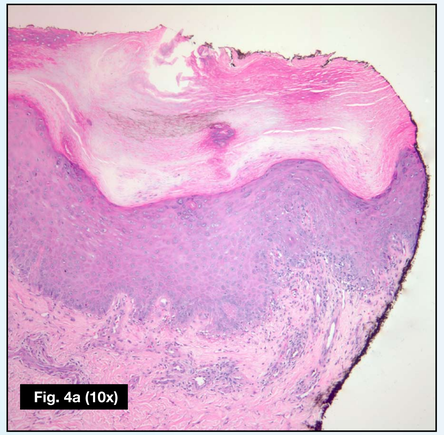

Figure 4a: Magnification 10x- Mild acanthosis and patchy parakeratosis with sparse perivascular infiltrate.

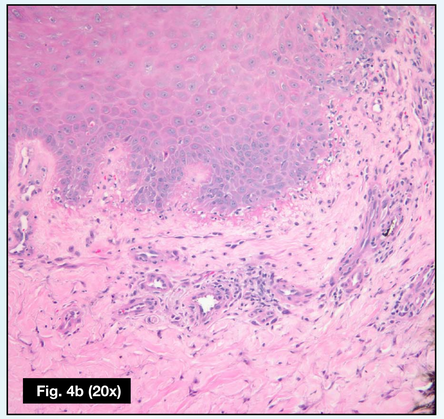

Figure 4b: Magnification 20x.

Discussion:

Dermatomyositis (DM) is an autoimmune systemic disease that can involve the skin, musculoskeletal, gastrointestinal, cardiac and pulmonary systems. Pathognomonic cutaneous features include the heliotrope rash and Gottron's papules. Gottron's papules are flat-topped erythematous papules and plaques typically found on joints of the hands and elbows. Poikiloderma, malar erythema, periungual telangiectasia, and photosensitivity are other common characteristic findings. Dystrophic and ragged cuticles, known as Samitz sign, can be observed. Proximal muscle weakness may occur before, during, or after the presence of cutaneous findings. Systemic manifestations can present as arthralgia, arthritis, dyspnea, dysphagia, arrhythmia, and dysphonia.

Laboratory studies may yield an elevated creatine kinase, aldolase, aspartate aminotransferase or lactic dehydrogenase due to myositis. A positive antinuclear antibody result is common but not diagnostic. Several myositis-specific antibodies have been identified but are not routinely used in diagnosis. They may, however, aid in the classification of DM subtypes for prognosis. Anti-Mi-2 antibody is highly specific and is associated with acute-onset classic DM with a relatively good prognosis. Anti–Jo-1 antibodies are more common in patients with polymyositis and can be associated with interstitial lung disease, Raynaud phenomenon, and arthritis. Juvenile onset DM has the best prognosis for survival, while paraneoplastic DM has the worst prognosis. A malignancy work up should be performed in all patients with adult onset DM. Multiple associated malignancies have been reported, including ovarian and breast cancer in women and lung cancer in men.

Erythroderma is an uncommon presentation of DM and can be associated with an underlying malignancy. Documented cases include erythrodermic DM associated with gastric cancer, liver cancer, and adenocarcinoma. Our patient with erythrodermic DM has had a negative malignancy work up to date. After 3 months on mycophenolate mofetil, hydroxychloroquine, prednisone, and topical clobetasol, he has cleared significantly. He occasionally experiences musculoskeletal pain but no longer experiences weakness.

Laboratory studies, malignancy work up, referral to appropriate specialists, and treatment to control cutaneous and muscle disease are important in the management of DM. Treatment options include topical and systemic corticosteroids, antimalarial agents, and immunomodulators. Systemic corticosteroids has been first line in the treatment of muscle disease. Methotrexate, mycophenolate mofetil, or azathioprine can be used as steroid-sparing agents. Sun avoidance and sun protection are important to avoid further exacerbations of skin disease.

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REFERENCES:

1 Iorizzo LJ 3rd, Jorizzo JL. The treatment and prognosis of dermatomyositis: an updated review. *J Am Acad Dermatol.* Jul 2008;59(1):99-112.

2 Kim SW, Kang YS, Park SH, et al. A case of Erythrodermic Dermatomyositis Associated with Gastric Cancer. *Ann Dermatol* 2009;21(4):435-9.

3 Nousari H, Kimyai-Asadi A, Spegman D. Paraneoplastic dermatomyositis presenting as erythroderma. *J Am Acad Dermatol* 1998;39:653-4.

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